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Staged Bilateral Total Hip Arthroplasty in a 17-Year-Old With Type VI Mucopolysaccharidosis

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ABSTRACT

Mucopolysaccharidosis encompasses multiple lysosomal storage disorders that are relevant to the orthopedic surgeon as they lead to disruption in bone and cartilage development. These patients may present with early-onset joint pain, including end-stage hip arthritis warranting total hip replacement. The altered hip anatomy in this disorder is of specific importance to the arthroplasty surgeon as it presents challenges when reconstructing the proximal femur and acetabulum and informs implant choice. We present a 17-year-old patient with end-stage bilateral hip arthritis who underwent staged bilateral total hip arthroplasty. We discuss technical considerations in surgical technique and the consequences of acetabular and femoral deformity on implant selection.

KEY POINTS

- It is important to understand the altered hip anatomy in these patients as the ideal cup and stem position is more challenging given their short stature, altered spinopelvic relationship, and both acetabular and proximal femur dysplasia.
- We suggest meticulous preoperative planning, including full-length lower-extremity radiographs and computed tomography scan, to assess optimal component position and bone stock.
- Regarding the equipment to have available in the operating room, we recommend having small acetabular shells, augments, and small diaphyseal engaging stems capable of adjusting anteversion and possible modular options to address leg-length discrepancies.

Introduction

Mucopolysaccharidosis (MPS) is a group of 13 lysosomal storage disorders that manifest in progressive multiorgan system dysfunction, including debilitating conditions of the musculoskeletal system. MPS is characterized by the deficiency of glycosaminoglycan (GAG)-degrading enzymes, leading to the accumulation of GAGs within lysosomes [1]. GAGs are viscous polysaccharides that couple with proteins to form proteoglycans and play a critical role as adhesion molecules. In articular cartilage, GAGs form complexes with water giving it compressive strength. GAGs are also prevalent in hyaluronic acid and the connective tissues throughout the body. In MPS, GAG buildup in lysosomes leads to cellular dysfunction and tissue damage [2]. The abnormal metabolism of GAG-containing proteoglycans further disrupts bone and cartilage development via interactions with growth factors including bone morphogenic proteins, fibroblast growth factors, and the Wnt signaling pathway [2–4].

There are multiple musculoskeletal manifestations seen in MPS including craniocervical stenosis, C1-C2 instability, kyphoscoliosis, abnormal epiphyseal formation, genu valgum, short stature, and hypermobility [4,5]. Early-onset debilitating joint pain is common in MPS, with the hip being one of the most commonly affected joints to develop end-stage arthritis [6]. Classically, the hip anatomy associated with MPS includes a dysplastic acetabulum, valgus femoral neck, and irregular ossification or resorption of the superomedial femoral head [7,8]. Given the amount of pain and loss

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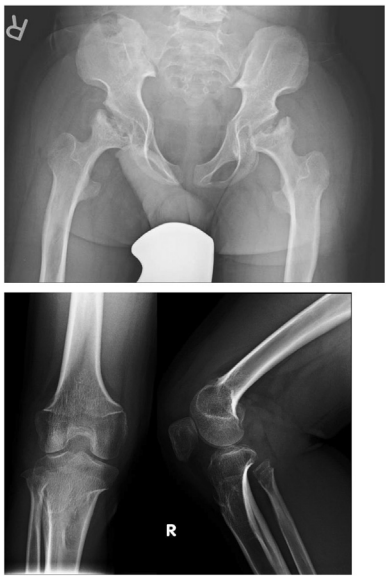

Figure 1. Initial anteroposterior (AP) pelvis and right knee radiographs.

of function, these patients are often candidates for total hip arthroplasty (THA). Anatomic features of MPS, such as shortened long bones, hip flexion contractures, and increased soft-tissue laxity present challenges to orthopedic surgeons tasked with joint reconstruction in these patients. The patient was informed that his clinical information would be submitted for publication and provided consent.

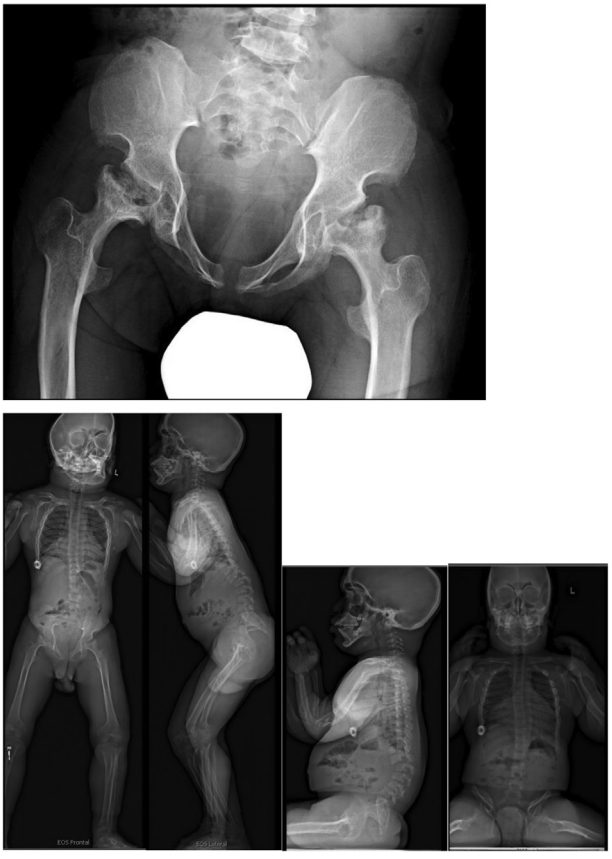

Figure 2. AP pelvis and standing and sitting radiographs.

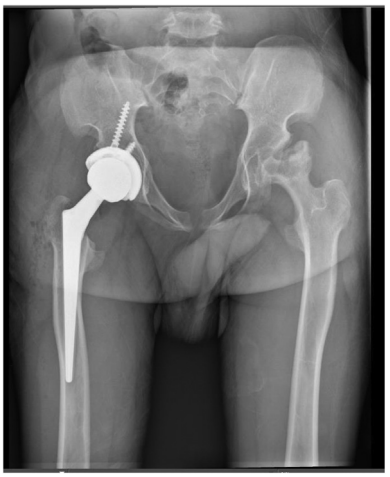

Figure 3. Postoperative right THA AP pelvis radiograph.

Case history

A 17-year-old male (weight, 36 kg; height, 121 cm; body mass index, 25.0 kg/m^2) with MPS type IVA presented to the clinic with 3 years of bilateral hip pain and loss of function. The patient initially presented to the clinic 15 months prior with right hip and right knee pain (Fig. 1). At that time, the patient was limited to ambulating 1 block with no assistive devices due to pain and sensation of unsteadiness on his feet. Otherwise, he used a motorized wheelchair. At that time, he was referred to musculoskeletal radiology for an image-guided right hip corticosteroid injection. This provided 1 week of symptom relief of both the hip and knee pain.

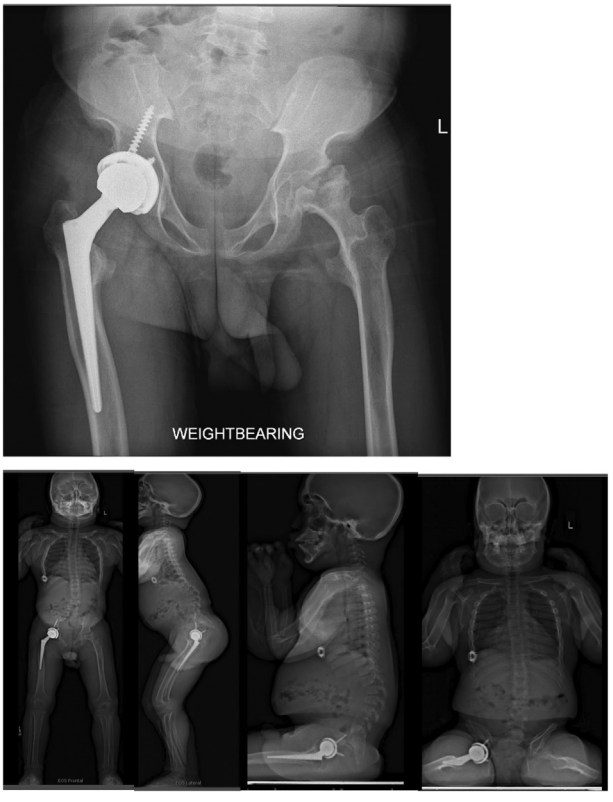

Figure 4. Postoperative radiograph 5 months from the right THA.

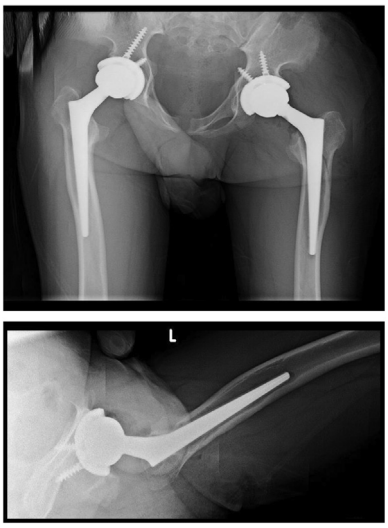

Figure 5. Postoperative left THA AP pelvis and cross-table lateral of left hip radiographs.

At presentation, the patient was limited to household ambulation and required a rolling walker. His right knee pain was predominantly retropatellar. On physical examination, his right hip was notable for a positive Stinchfield test, 90 degrees of passive hip flexion, and 0 degree of internal or external rotation. Given his failure to improve with nonoperative management including physical therapy and corticosteroid injection, as well as rapid loss of function, the patient was indicated for right THA (Fig. 2).

Surgery was performed via a posterior approach with the aid of computer navigation (Intellijoint HIP; Waterloo, Canada). A 44-millimeter (mm) acetabular shell (Stryker, Kalamazoo, MI) and 35-mm and 25-mm screws were placed. An uncemented, 14-mm 125-degree monoblock tapered and fluted stem was used (Wagner Cone; Zimmer-Biomet, Warsaw, IN). The bearing surface was neutral-faced highly crossed polyethylene liner and a 32-mm ceramic head (Zimmer-Biomet, Warsaw, IN) (Fig. 3).

Postoperatively, the patient recovered well. His pain improved, and he was able to ambulate comfortably with a rolling walker. At 3 months postoperatively, he began to complain of increased progressive left hip pain. As he was pleased with the result of the right hip, he was indicated for left THA (Fig. 4).

Approximately 8 months after undergoing the right THA, the patient underwent a left THA. Besides using a +7-mm head, all components matched the contralateral side. The patient was discharged home the morning of postoperative day 1 (Fig. 5).

Currently, the patient is doing well 7 months after the left THA and 11 months after the right THA. He reports no hip pain and is ambulating with a rolling walker (Fig. 6).

Discussion

In this case, our patient has MPS type IV, also known as "Morquio syndrome." This form of MPS affects 1 in 200,000 and is characterized by an accumulation of keratan sulfate. There are two sub-types of MPS IV; type IVA involves galactosamine-6-sulfatase deficiency, and IVB involves a beta-galactosidase deficiency. While understanding the metabolic intricacies of this disorder may not be crucial for the orthopedic surgeon treating hip pathology in these patients, there are specific features associated with this disorder that surgeons should consider. In these patients, growth typically stops by 8 years of age, patients on average live into the third or fourth decade of life, and some live to be much older [9]. The hip degeneration in MPS type IVA is an issue of biologic as well as mechanical factors. While enzymatic deficiency causes developmental abnormalities in endochondral ossification and articular cartilage, there is also concomitant acetabular dysplasia leading to accelerated deterioration of the hip joint [10].

With regard to the technical aspects of the hip reconstruction, these patients present several challenges. In the preoperative assessment, it is important to obtain full-length standing and sitting radiographs as it is crucial for the preoperative plan in these patients. Classically, these patients have a focal thoracic kyphosis with compensatory hip flexion contractures. This is important to consider when optimizing cup position as hip flexion contractures and spinopelvic motion can improve after THA (as seen in Fig. 7). In addition to aiding in understanding the functional position of the pelvis, full-length views will help recognize leg length discrepancies as well as any extra-articular deformity of the femur.

Recognizing the severity of the acetabular deformity is crucial in these case as these patients tend to have varying degrees of hip dysplasia. Depending on the severity, a computed tomography scan may be necessary to better assess the native femoral anteversion, position of their native acetabulum, and the available acetabular bone stock. Dysplastic acetabuli tend to be shallow, ovoid, and retroverted. It is important to recognize the acetabular morphology

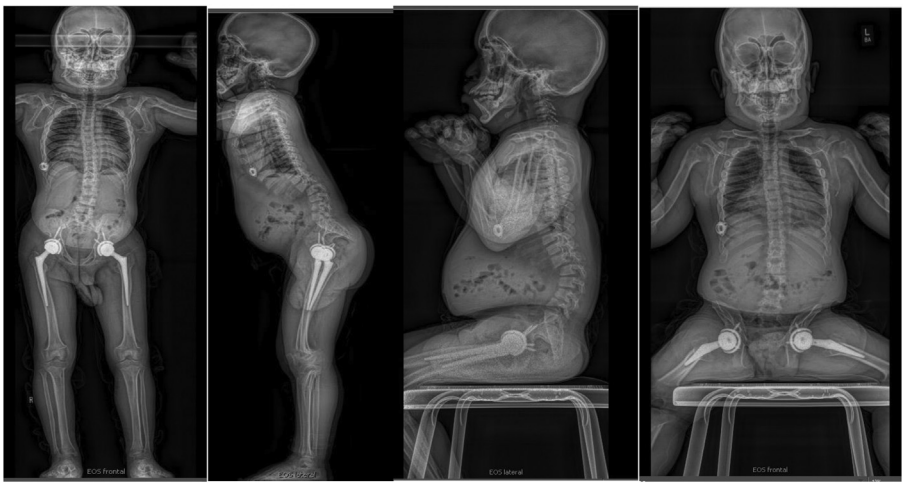

Figure 6. Postoperative radiograph 3 months from the left THA.

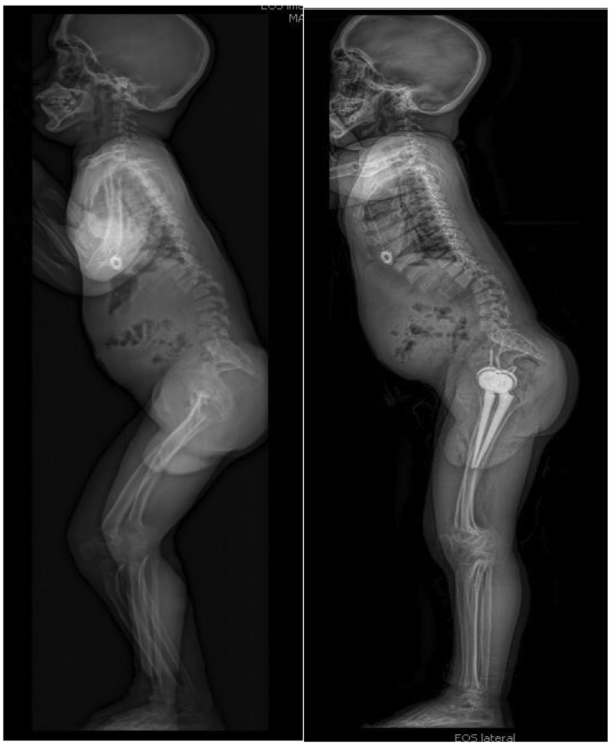

Figure 7. Standing and sitting lateral radiographs performed 3 months after the left THA.

to plan the ideal cup position. Furthermore, for these patients, it is important to have small acetabular shells and acetabular augments available. Additionally, the use of a thinner polyethylene liner is an option to maximize head size to confer more stability.

In regard to the proximal femur's anatomy, patients with MPS tend to have smaller and shorter femoral canals with anteverted, valgus femoral necks. This poses a challenge with femoral component options. With this femoral geometry, a cylindrical metaphyseal/diaphyseal-engaging stem is preferred for many, which are typically ream-only systems. While the implant in this case is a monoblock design, there are several diaphyseal engaging, modular stems. Modular designs with a proximal body allow adjustment of leg length, in addition to the ability to dial in the femoral version as seen in monoblock designs.

For cases such as the one presented here, it is imperative to have smaller implant sizes available, as they are not commonly used sizes at hospitals or surgery centers. Furthermore, knowing the head options available with these smaller acetabular components prior to the case is an important consideration in planning for these cases effectively.

In addition to the anatomic challenges of the case, stem fixation is an important consideration. For most surgeons, cementless fixation is likely preferable in a young otherwise healthy patient with good bone quality as it may provide long-term biologic fixation. However, in patients with compromised bone quality or femoral geometry that is not accommodating of a cementless prosthesis, a cemented option is a technique shown to provide good long-term results.

Summary

THA can be very successful in the treatment of end-stage hip arthritis for patients with MPS. We present a 17-year-old patient with bilateral hip arthritis treated with staged bilateral THA. It is important to understand the altered hip anatomy in these patients as the ideal cup and stem position is more challenging given their short stature, altered spinopelvic relationship, and both acetabular and proximal femur dysplasia. We suggest meticulous preoperative planning, including full-length lower-extremity radiographs and computed tomography scan, to assess optimal component position and bone stock. In regard to equipment available in the operating room, we recommend having small acetabular shells, augments, and small diaphyseal engaging stems capable of adjusting anteversion and possible modular options to address leg-length discrepancies.

Conflicts of interest

R.S. is a paid consultant for Smith & Nephew and Intellijoint. He also has stock options in Gauss Surgical and PSI outside the submitted work. J.X.R. and A.I.B. have nothing to disclose. All authors were not involved in the journal's review of or decisions related to this manuscript.

Informed patient consent

The author(s) confirm that written informed consent has been obtained from the involved patient(s) or if appropriate from the parent, guardian, power of attorney of the involved patient(s); and, they have given approval for this information to be published in this case report (series).

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